

# Transitions and touchpoints in idiopathic pulmonary fibrosis

Jeffrey J Swigris

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## ABSTRACT

Patients with idiopathic pulmonary fibrosis (IPF) face a poor prognosis and endure intrusive symptoms that impair quality of life. Many patients with IPF will require supplemental oxygen (O<sub>2</sub>) at some point in the course of their illness, and although it can improve blood oxygen and symptoms, O<sub>2</sub> creates physical and emotional challenges for patients and their loved ones. Four events in the course of IPF—the first occurs at the time of diagnosis and the other three are related to O<sub>2</sub>—herald periods of transition for patients and their caregivers and mark touchpoints when they need extra care and support from practitioners.

touchpoint: a time, condition, or circumstance that is vulnerable or unstable enough to precipitate a highly unfavourable, possibly devastating outcome

– English Oxford Dictionary

## INTRODUCTION

Idiopathic pulmonary fibrosis (IPF) is a typically life-shortening condition whose debilitating symptoms include activity-limiting dyspnoea, nagging cough and fatigue.<sup>1</sup> At the time they are diagnosed with IPF, patients (and their informal caregivers) are forced to confront the ‘devastation’ and begin thinking about a life with this disease.<sup>2</sup>

Most patients with IPF will require supplemental oxygen (O<sub>2</sub>) at some point after diagnosis. Although O<sub>2</sub> is prescribed in the hopes of preventing long-term consequences of hypoxaemia and improving symptoms, it poses a threat to already-impaired quality of life (QOL) in these patients<sup>3</sup>: many feel stigmatised when they are seen in public wearing a nasal cannula, and using O<sub>2</sub> robs patients of their ability to live as carefree and independently as they would like.<sup>3 4</sup> Understandably, patients view the need for O<sub>2</sub> as a marker of disease progression—an unwanted milestone in the course of IPF that, like being told the diagnosis, can cause their worlds to ‘crash for a second time.’<sup>5</sup> Compared with patients with IPF who do not need O<sub>2</sub>, those who need it report worse QOL in multiple life domains,

including emotional well-being, social participation and independence.<sup>6</sup> Patients with IPF are resilient: in clinic, I frequently hear patients with IPF say things like, ‘I can deal with this... if it just doesn’t get any worse’. But, it so often does—and they deal with it anyway and figure out how to move on.

The closest thing to a formal staging system for IPF is the GAP index (a combination of gender, age and physiological variables), which can be generally informative in discussions of prognosis.<sup>7</sup> But patients with IPF also want information on what life is going to look like with IPF—how disease progression is likely to affect them in their daily lives and what things they should be doing or thinking about along the way. Informal caregivers want similar information about the patient. They also want to know how their roles will change as disease severity increases, and they need to know that helpful resources are accessible to them.

I believe care of patients with IPF requires an ongoing conversation (with them and their caregivers) woven through serial visits over the course of the disease. This conversation forms the backbone of the palliative care these patients need—unfortunately, the term ‘palliative care’ has become synonymous with end-of-life care, and perhaps more appropriate terms to use are ‘comprehensive care’ or ‘quality of life (QOL) care’. Regardless, the conversation should, among other topics personalised to a given patient, include an assessment of disease status, an appraisal of the tolerance of and response to therapeutic manoeuvres (pharmacological and/or non-pharmacological), an evaluation of the patient’s and the caregiver’s physical and emotional well-being, assurance that necessary supportive resources are in place, and time for questions. As with any patient with a potentially life-shortening condition, this longitudinal conversation must involve appropriately timed talk around advanced care planning and, most importantly, patients’ wishes for end-of-life intervention and care.<sup>5 8 9</sup> Like other topics in the weave,



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Interstitial Lung Disease Program, National Jewish Health, Denver, Colorado, USA

**Correspondence to**  
Dr Jeffrey J Swigris;  
swigrisj@njc.org

these are best brought up early, before the pressures and chaos of abrupt decline mandate. Here, practitioners have the challenging duty of (1) ascertaining what information patients and caregivers are ready to receive, (2) recognising that patients' and caregivers' levels of acceptance—or as Overgaard and colleagues called it, 'reactional stage'<sup>5</sup>—may differ, and thus (3) carefully tailoring information delivery to the recipient while considering their readiness to receive it.

I view four major events in the course of IPF as meriting special care and attention and forming a framework for the disease-long conversation. The first is when the diagnosis is given. Because of the potential impact of O<sub>2</sub>,<sup>2-6</sup> the final three events centre on it: when O<sub>2</sub> is prescribed for use with exertion; when O<sub>2</sub> is needed at rest; when high-flow O<sub>2</sub> is required (ie, when a portable oxygen concentrator or other lightweight, portable, O<sub>2</sub> conserving device does not meet a patient's oxygen demands).

I initially regarded these events as stages because, to some degree, they often correspond with disease severity. But on reflection, they are much more: for patients and caregivers, both individually and for them as a team, these events are times of transition, when each is particularly vulnerable, when their QOL is most threatened, and they are forced to adapt to a new normal. More than perhaps any other times in the course of IPF, these are the occasions when patients and caregivers need practitioners as partners—accessible, ready to give information, offer support, bolster realistic hope and attend to their emotional well-being and QOL. For practitioners, these events can serve as touchpoints—special opportunities to intervene when we have the chance to more fully understand patients' and caregivers' preferences, values and goals through comprehensive needs assessments. Whether done systematically using a tool specifically developed for this purpose<sup>10-12</sup> or spontaneously in the natural flow of a visit, the assessments are critical to QOL care. Because patients' and caregivers' needs change over time, the touchpoints can serve as both reminders and opportunities for the assessments to occur.

## WHEN THE DIAGNOSIS IS GIVEN

### Transition

Whether a patient presents knowing that something serious is going on, or thinking they are just overweight, out of shape and 'old', being told they have IPF will likely turn their lives upside down.<sup>13</sup> They become contemplative, and many experience grief, worrying about their (and their family's) futures as they must 'refocus their lives' and 'readjust their life goals'.<sup>13</sup> For many patients with IPF, not dwelling on the negative is challenging, but over time, by gaining knowledge and feeling supported, most adjust and accept life with IPF. Caregivers experience similar grieving, disbelief and worry—wondering how they will carry on if their patient loved-one lives only as long as the average patient with IPF. The transition to

acceptance—both the time and route—is different for every patient and caregiver.<sup>5</sup>

### Touchpoint

Those of us who diagnose IPF and care for patients with it know the devastation our words can bring when we tell someone they have IPF. Doing so with care and empathy softens the blow, but patients' and their caregivers' lives are forever changed. Making certain that our words convey realistic hope, taking time to educate patients and their caregivers about the condition, informing them that there are therapies available for patients with IPF (pharmacological and non-pharmacological) and, together, developing a plan for moving forward are critical at this touchpoint. Handing out or directing patients to trustworthy disease education materials, informing them about local support groups and firming up plans for a follow-up visit in the near future are helpful.

## WHEN O<sub>2</sub> IS PRESCRIBED FOR USE WITH EXERTION

### Transition

For many patients with IPF, particularly those who do not need O<sub>2</sub> at the time of diagnosis, being told they now need it is another demoralising milestone—'a big step' in the wrong direction.<sup>2</sup> Some patients accept O<sub>2</sub> and figure out how to accommodate it immediately. Other patients do not accept it and decline the prescription; they prefer to slow down and/or cut out certain activities altogether rather than use O<sub>2</sub>. Still others navigate their way through stages of negotiation and compromise in which they decide how and when O<sub>2</sub> fits into the reality of their daily lives.<sup>14</sup> In the end, the patients who accept O<sub>2</sub> transition to a reluctant acceptance of it and find a way to exist in the new normal it creates.

### Touchpoint

The lack of robust data in support of O<sub>2</sub> for patients who desaturate only with exertion notwithstanding, scientific rationale and clinical intuition would suggest that intermittent hypoxaemia in people 60–80 years old with one or (typically) more chronic medical conditions could have detrimental effects. Limited published data show that O<sub>2</sub> increases activity and/or decreases dyspnoea in patients with IPF who are normoxic at rest but desaturate with activity.<sup>15 16</sup> Additional studies whose results will be published soon should shed more light on the topic.<sup>17 18</sup> Regardless of how the data are interpreted or whether patients agree to use O<sub>2</sub>, when peripheral oxygen desaturation is identified the first time, the conversation changes. A thoughtful, transparent discussion—one that involves attentive listening to patients' and caregivers' concerns, a review of expectations and the potential challenges O<sub>2</sub> poses—promotes informed, shared decision-making that prominently incorporates their values and preferences. *Adventures of an Oxyphile* by Thomas L Petty and coauthors (published by Snowdrift Pulmonary

Conference) is a great resource that I direct my patients and their caregivers to at this touchpoint. It offers hope by showing that life does not end when O<sub>2</sub> is needed.

### WHEN O<sub>2</sub> IS NEEDED AT REST

#### Transition

When patients transition to needing O<sub>2</sub> at rest, they really feel its constraints. The stationary concentrator that sat in the bedroom, used only during sleep, may have to be moved; the 7-foot cannula will have to be lengthened considerably, so patients can move throughout the home. With all the planning O<sub>2</sub> requires—the tank filling, the deliveries from the supplier, the equipment and tubing—patients and caregivers begin to feel like their lives revolve around O<sub>2</sub>. And, once again, they are thrust onto a new, lower plateau of independence and freedom. In an interesting paradox, some caregivers, for the first time in the course of their loved-one's disease, perceive themselves as truly useful at this transition: instead of just being a 'nag', caregivers can give physical care—helping with the logistics of O<sub>2</sub> and taking on more household duties.<sup>9</sup>

#### Touchpoint

As a partner to patients and their caregivers, practitioners have a lot to offer at this touchpoint—being a source of empathic positivity is key. This is another time when patients need to be reminded that there is more living to be done. Patients and caregivers should be encouraged to focus on what they can do, not what they cannot. Referring (or re-referring) to pulmonary rehabilitation is an extremely useful intervention to reinforce to patients and caregivers the possibility and importance of remaining active.

This is also a time when a discussion of alternative O<sub>2</sub> delivery modalities is reasonable. For many patients with IPF, delivery of O<sub>2</sub> via the transtracheal route (TTO) is an attractive option. The benefits of TTO are that it gets the cannula out of the nose, and on average, it allows reduction of O<sub>2</sub> flow by 30%–50%.<sup>19</sup> TTO catheters require daily care; manual dexterity is required to clean and reinsert them, and some patients just do not like the idea of having 'a hole in my neck'. In our experience, TTO catheters are most beneficial when inserted before O<sub>2</sub> requirements reach 6 L/min. Another option for O<sub>2</sub> delivery is via Oxy-View glasses (Oxy-View, Englewood, Colorado, USA): the supply tubing connects to the ends of the temple bars, and O<sub>2</sub> is delivered through the hollow frame to small, unobtrusive cannulas that run down the side of the nose in the alarfacial groove to enter the nostril. These are particularly attractive for patients who also need glasses and hearing aids. Patients must be reminded that an O<sub>2</sub> source is still necessary.

Ideally, discussions about lung transplantation (and potentially referral to a transplant programme) have taken place, but if not, this is a topic that should be added to the conversation at this touchpoint. Although

robust data are lacking, pharmacological intervention for dyspnoea (eg, opiates) is an option that could improve symptoms and/or exercise capacity in some patients.

### WHEN HIGH-FLOW O<sub>2</sub> IS NEEDED

#### Transition

Newer technologies are in development, but currently, lightweight, portable devices deliver only so much O<sub>2</sub>—and only for so long. When these devices are unable to meet their needs, patients typically see two options: (1) cart around larger tanks or (2) exert less. At this transition, patients and their caregivers feel more 'tethered' to home than ever before—and it takes a special determination to break through the constraints.

#### Touchpoint

Those of us who practise at higher elevations may care for more patients who make this transition than practitioners in other places; however, all patients in the latter stages of IPF (or those with coexisting emphysema or pulmonary hypertension) are at risk. Particularly for patients who remain active and desire to leave the home, by increasing the amount of O<sub>2</sub> they can carry, liquid O<sub>2</sub> can alleviate substantial burden for patients in this transition. An E (M-24) tank holds 680 L of gaseous oxygen; 1 L of liquid O<sub>2</sub> is equivalent to 860 L of gaseous O<sub>2</sub>. A commonly used, larger, portable, liquid O<sub>2</sub> unit holds 1058 L gaseous O<sub>2</sub> equivalents at the same weight as an E tank. Unfortunately, liquid O<sub>2</sub> is being phased out in many areas across the USA. An option for home that I have yet to prescribe for my patients—but one that I am enthusiastic about trying—is humidified and heated nasal high-flow O<sub>2</sub> delivered via a device like the myAIRVO2 (Fisher and Paykel Healthcare, Auckland, New Zealand).

At this touchpoint, another reminder that high-flow O<sub>2</sub> is not the end of the road can be powerful. Some patients may need encouragement to try to stay physically and socially active, but we should remember that many will grieve the loss of their 'former' lives and the ability to do some of the activities they once enjoyed—or at least the ability to do them with the gusto they once did. Sharing examples of other patients who have found new activities or ways to continue to do the things they always enjoyed can give hope to those in this transition.

### CONCLUSION

IPF is a serious condition that steals life's length and quality from patients and their caregivers. Despite the potentially dire outlook and limited therapeutic interventions available, there is much for practitioners to offer these people: information, realistic hope and a plan—partnership throughout the course of the condition. The cornerstones of this partnership are accessibility and a longitudinal conversation. In the course of IPF, we practitioners need to be on high alert for the appearance of

four life-changing transitions when patients and their caregivers are most vulnerable and their QOL is most threatened. Practitioners can use these high-alert times as touchpoints to affirm the partnership they have established with the patient, to assess patients' and caregivers' needs and well-being, and to solidify plans that focus on patient and caregiver QOL no matter what the future holds for them.

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## REFERENCES

1. Raghu G, Rochweg B, Zhang Y, *et al*. An official ATS/ERS/JRS/ALAT clinical practice guideline: treatment of idiopathic pulmonary fibrosis. an update of the 2011 clinical practice guideline. *Am J Respir Crit Care Med* 2015;192:e3–e19.
2. Belkin A, Albright K, Swigris JJ. A qualitative study of informal caregivers' perspectives on the effects of idiopathic pulmonary fibrosis. *BMJ Open Respir Res* 2014;1:e000007.
3. Cullen DL, Stiffler D. Long-term oxygen therapy: review from the patients' perspective. *Chron Respir Dis* 2009;6:141–7.
4. Khor YH, Goh NSL, McDonald CF, *et al*. Oxygen therapy for interstitial lung disease. A mismatch between patient expectations and experiences. *Ann Am Thorac Soc* 2017;14:888–95.
5. Overgaard D, Kaldan G, Marsaa K, *et al*. The lived experience with idiopathic pulmonary fibrosis: a qualitative study. *Eur Respir J* 2016;47.
6. Swigris JJ, Wilson SR, Green KE, *et al*. Development of the ATAQ-IPF: a tool to assess quality of life in IPF. *Health Qual Life Outcomes* 2010;8:77.
7. Ley B, Ryerson CJ, Vittinghoff E, *et al*. A multidimensional index and staging system for idiopathic pulmonary fibrosis. *Ann Intern Med* 2012;156:684–91.
8. Bajwah S, Higginson IJ, Ross JR, *et al*. The palliative care needs for fibrotic interstitial lung disease: a qualitative study of patients, informal caregivers and health professionals. *Palliat Med* 2013;27:869–76.
9. Ramadurai D, Corder S, Churney T, *et al*. Understanding the informational needs of patients with IPF and their caregivers: 'You get diagnosed, and you ask this question right away, what does this mean?'. *BMJ Open Qual* 2018;7:e000207.
10. Boland JW, Reigada C, Yorke J, *et al*. The adaptation, face, and content validation of a needs assessment tool: progressive disease for people with interstitial lung disease. *J Palliat Med* 2016;19:549–55.
11. Johnson MJ, Jamali A, Ross J, *et al*. Psychometric validation of the needs assessment tool: progressive disease in interstitial lung disease. *Thorax* 2017;thoraxjnl-2017-210911.
12. Reigada C, Papadopoulou A, Boland JW, *et al*. Implementation of the Needs Assessment Tool for patients with interstitial lung disease (NAT:ILD): facilitators and barriers. *Thorax* 2017;72:1049–51.
13. Swigris JJ, Stewart AL, Gould MK, *et al*. Patients' perspectives on how idiopathic pulmonary fibrosis affects the quality of their lives. *Health Qual Life Outcomes* 2005;3:61.
14. Earnest MA. Explaining adherence to supplemental oxygen therapy: the patient's perspective. *J Gen Intern Med* 2002;17:749–55.
15. Frank RC, Hicks S, Duck AM, *et al*. Ambulatory oxygen in idiopathic pulmonary fibrosis: of what benefit? *Eur Respir J* 2012;40:269–70.
16. Visca D, Montgomery A, de Lauretis A, *et al*. Ambulatory oxygen in interstitial lung disease. *Eur Respir J* 2011;38:987–90.
17. Belkin A, Fier K, Albright K, *et al*. Protocol for a mixed-methods study of supplemental oxygen in pulmonary fibrosis. *BMC Pulm Med* 2014;14:169.
18. Visca D, Tsipouri V, Mori L, *et al*. Ambulatory oxygen in fibrotic lung disease (AmbOx): study protocol for a randomised controlled trial. *Trials* 2017;18:201.
19. Christopher KL, Schwartz MD. Transtracheal oxygen therapy. *Chest* 2011;139:435–40.